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Case Report

Vagal nerve schwannoma: a rare case from Border Security Force Composite hospital

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ABSTRACT

Cervical vagal schwannoma is an extremely rare entity, slow growing, solitary, asymptomatic benign neck mass associated with vagus nerve. Definitive pre-operative diagnosis may be difficult and investigations such as fine needle aspiration biopsy (FNAB) have low specificity. Surgical excision is the treatment of choice. We describe a 26-year-old female presenting as right neck mass who underwent surgery. Magnetic resonance imaging was done, and patient was operated under general anaesthesia. Intraoperatively, the tumour was found to be originating from the vagus nerve. Final histopathology confirmed our preoperative suspicion of vagal nerve schwannoma. Vagus nerve schwannomas should be distinguished from other tumours that arise in the neck before planning surgery, to minimize the risk of nerve injury. A careful surgical technique is mandatory for tumour clearance and to reduce postoperative complications.

Keywords: Magnetic resonance imaging, Schwannoma, Vagus nerve

INTRODUCTION

A schwannoma, also known as an acoustic neuroma, is a benign nerve sheath tumour of schwann cells, which normally produce the insulating myelin sheath covering the peripheral nerves. Schwannoma, originating from the cervical vagus nerve, is an extremely rare neoplasm that usually occurs in men between 3rd and 6th decade of life and preoperative diagnosis is generally difficult, since most cervical vagal schwannomas do not present with neurological deficits. Paragangliomas, branchial cleft cysts, malignant neck tumours and cervical lymphadenopathy should be kept in mind in differential diagnosis.[1] As there are no specific imaging or histological features, a great deal of clinical suspicion is needed to make an accurate preoperative diagnosis.[2] Investigations like FNAC have low specificity. Magnetic resonance imaging has become the routine imaging study for these tumours. Histologically, the tumour is composed of bundles of spindle cells, which are strongly positive for S-100 protein detected using immunohistochemistry. We hereby present a rare case of cervical vagal nerve schwannoma presenting as a huge mass in the right carotid apace, who underwent surgery.

CASE REPORT

A 26-year-old female patient was admitted to our hospital with complaints of painless mass over the right cervical region of more than 6month duration. On clinical examination, a mass was found in the right level 2 cervical region which was 7cmx4cm in diameter, smooth surfaced, mobile in horizontal plane, firm and non-tender swelling. There was no pulsation or murmur heard over the mass. A contrast enhanced computed tomography revealed a heterogeneously enhancing mixed density space occupying lesion in right lateral triangle of neck with internal areas of calcification and cystic areas of

necrosis causing extrinsic compression and displacement of the great vessels of neck. Magnetic resonance imaging of the neck confirmed a well-defined mass measuring 5cm x 4cm x 6.6cm in right carotid space extending from C 1-2 to C 5-6 level. The mass was causing displacement of the internal carotid artery, external carotid artery and common carotid artery anterolaterally and is abutting the anterior margin of the mass. The internal jugular vein was compressed and is draped along the anterio-lateral margin of the mass (Figure 1).

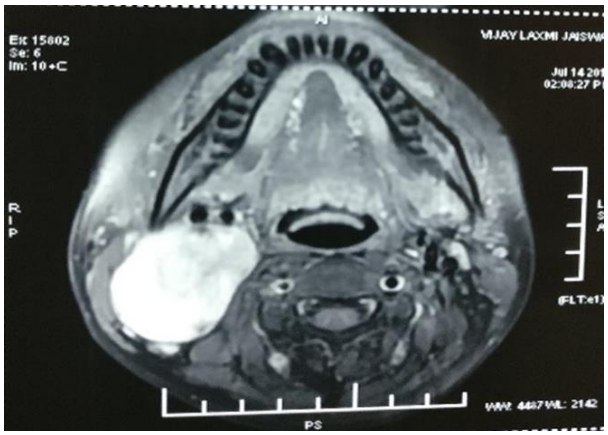

Figure 1: MRI showing the vagal nerve schwannoma compressing the major vessels of neck.

Fine needle aspiration biopsy of the mass was attempted twice as it was missed initially but the second time revealed features consistent with benign nerve sheath tumour. Under general anaesthesia, an oblique cervical incision was made starting from the right mastoid apex extending upto the inferior border of mandible. A yellowish mass lesion was observed which was 7cm x 4cm in diameter originating from the vagus nerve and medially adjacent to the common carotid artery (Figure 2).

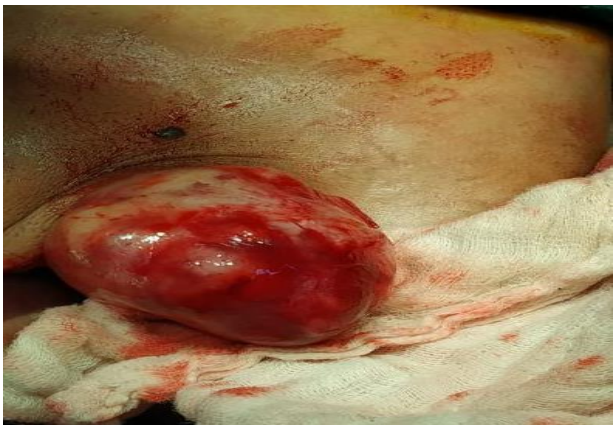

Figure 2: Intraoperative pic of vagal nerve schwannoma.

The mass was carefully dissected from the vagus nerve and other adjacent structure with care taken to protect the vagal nerve's integrity. The specimen was sent for histopathological examination. Postoperative course was uneventful.

On histopathological examination, a well encapsulated tumour of hypercellular Antoni A areas with Verocay bodies surrounded by Antoni B areas composed of hypocellular myxoid matrix with occasional cystic changes was seen, with features consistent with schwannoma (Figure 3).

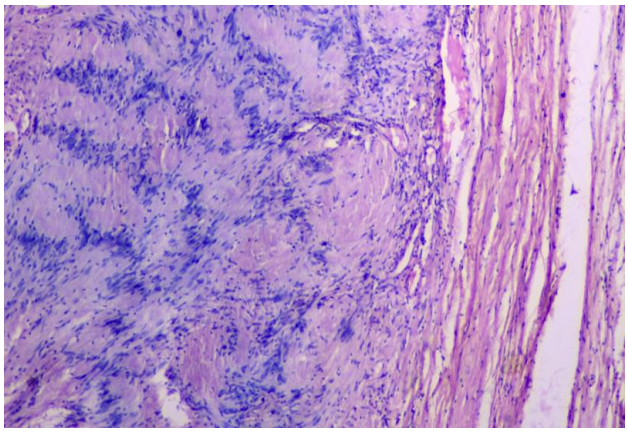

Figure 3: Histopathology of vagal nerve schwannoma.

DISCUSSION

Approximately 25-45% of extracranial schwannomas are present in the head and neck area; the most commonly affected region are the temporal bone, lateral neck and paranasal sinus.[3] The involvement of vagus nerve has been reported in 10% of cases. Most Schwannomas occur between the 3rd and 6th decade of life, affecting both the gender equally, and have minimal risk of malignant transformation. Schwannomas presents as a slow growing fixed and painless mass. When schwannoma arises in the neck, hoarseness of voice is the most common symptom.

However, a pathognomonic clinical sign is a cough on palpation of the mass. Diagnosing vagal schwannoma preoperatively may be difficult, since there are no classic imaging or cytological criteria to diagnose them. The classical histopathological picture is one composed of antony A bodies which signify compact spindle cells and antony B bodies which compromise looser arranged schwann cells.[4]

Adequate treatment involves complete removal of the lesion safeguarding the nerve involved. If it is impossible to find an adequate plane and is technically difficult to preserve the integrity of the nerve trunk, the involved segment may be resected and an end to end anastomosis performed using microsurgical techniques but may result in vocal nerve paralysis. Postoperative vocal cord palsy is a major concern in the surgical management. During the surgery, it is of paramount importance to have an attentive surgical technique with gentle and careful

dissection of the fascicles of the nerve and extreme caution to protect the adjacent structures is essential to avoid unwanted postoperative neural compromise.

Furukama et al, found that vagal schwannomas separate the common or internal carotid artery from the jugular vein, whereas schwannomas of the cervical sympathetic chain do not.[5] schwannomas have been commonly been associated with neurofibromatosis (NF) 1 and neurofibromatosis 2, but mostly NF 2. When multiple schwannomas are present in the absence of other stigmata of NF, then it can be defined as schwannomatosis.[6]

The reported incidence of preoperative vocal cord palsy is about 12% but postoperative hoarseness is almost present and therefore it is mandatory to access the vocal cord status prior to surgery[7]. Since vagal schwannomas are almost always benign, a conservative approach should always be considered in first instance when the integrity of the nerve is in question.[8] In the presence of post-operative vocal cord palsy, aggressive voice therapy, for vocal cord compensation, should be started soon after the surgery.[9] The prognosis of schwannomas after complete excision is good, and recurrence is rare.[10]

CONCLUSION

Though benign and slow growing tumour of the head and neck, schwannomas are rare and potentially morbid lesions. The preoperative diagnosis is mainly based on clinical suspicion. Complete resection of the tumour is the treatment of choice. However, a careful and meticulous surgical technique is mandatory for tumour clearance and to reduce postoperative complications.

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